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Case Report

Monostotic Fibrous Dysplasia of the Lumbar Spine: Case Report and Review of the Literature

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Summary: Fibrous dysplasia has been frequently reported to involve the spine in the polyostotic form, but only rarely has monostotic fibrous dysplasia been noted. In the only previously reported case involving the lumbar spine, the disease was confined to the transverse process. The present case demonstrates monostotic fibrous dysplasia involving the vertebral body in addition to the posterior elements. The plain radiographic, computerized tomography, and histologic examinations are presented.
Key Words: Lumbar spine—Monostotic fibrous dysplasia.

Fibrous dysplasia is a benign, relatively common fibroosseous lesion of bone. The earliest reports discussed only cases with involvement of multiple bones (1,10–12). Subsequently, three distinct forms of involvement were recognized (11,18) monostotic, polyostotic, and polyostotic with cutaneous and endocrine abnormalities (Albright syndrome) (1). The most frequent areas of involvement in each of these forms are ribs, proximal femur, tibia, skull, and maxilla (9,14,19,21). Spinal involvement is uncommon in the polyostotic form (10,11,14,20) and rare in the monostotic form (4,9,11,19,20).

CASE REPORT

The patient was referred for treatment of a lumbar lesion. She was a 12½-year-old white girl in good health until she fell from her bicycle. She sustained two phalangeal fractures and complained of midthoracic back pain. Radiographs of the spine demonstrated an expansile, lytic lesion of L3 (Fig. 1). A technetium bone scan showed increased uptake in the midlumbar region. Computerized tomography of the lumbar spine revealed a lytic, expansile lesion involving the right posterior body, pedicle, superior articular process, and right transverse process of L3 (Fig. 2). She was then referred for further evaluation.

On presentation to our institution 5 months after the initial trauma, the patient was asymptomatic. The medical history was significant for treatment as an infant for bilateral retinoblastoma with chemotherapy and local radiation followed by enucleation. At follow-up, she was considered to be disease free. Her general physical examination was within normal limits. She was premenarchal with no stigmata of endocrine abnormalities. The back was nontender and had a full range of motion. The

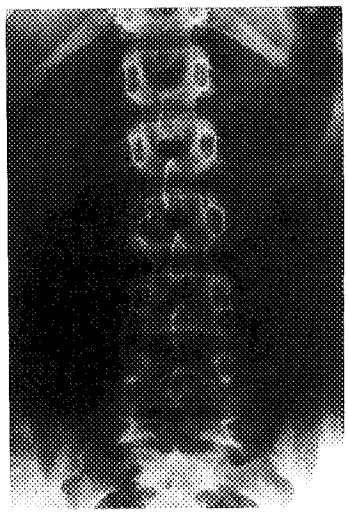

FIG. 1. Anteroposterior radiograph of the lumbar spine demonstrates an expansile, lytic lesion of L3 involving primarily the right posterior elements.

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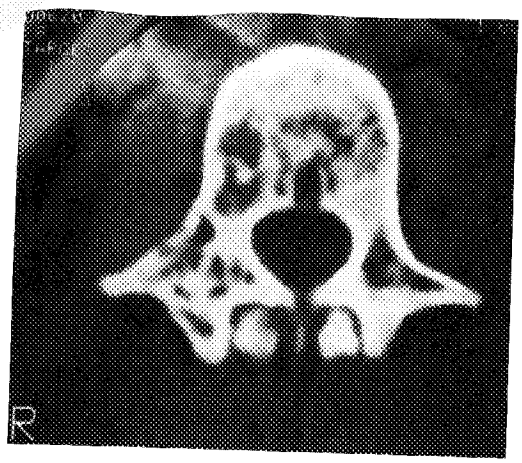

FIG. 2. Computerized tomography scan at the level of L3 demonstrates a lesion involving the right posterior body, right pedicle, right superior articular process, and right transverse process.

neurological examination was normal, and the initial laboratory examination was within normal limits. The presumptive diagnosis was aneurysmal bone cyst. Because of the history of malignancy as an infant and the atypical radiographic appearance of the lesion, a biopsy was performed.

Through a midline incision from L1 to the sacrum, the spine was exposed. The external appearance of the posterior elements was normal. The location was confirmed radiographically. The right superior facet and right superior portion of the transverse process of L3 were removed, and a cavity was entered. The cavity was surrounded by dense sclerotic bone with a thin membrane lining. There were sparse trabeculae transversing the cavity and no fluid. Following curettement of the lesion, autogenous bone graft obtained from the posterior iliac crests was packed in the cavity. A posterior fusion from L2 to L3 was performed. Microscopic examination showed woven bone with intervening fibrous tissue (Fig. 3). The pathological diagnosis was fibrous dysplasia.

The postoperative course was unremarkable. Before discharge, the patient was fitted with a posterior brace with anterior corset. The brace was removed at 11 weeks, at which time there was radiographic evidence of fusion (Fig. 4). When last seen 3 years after her initial injury, she was pain free with no scoliosis.

DISCUSSION

The earliest reports of fibrous dysplasia were primarily of severe polyostotic cases, most with the associated pigmentation and endocrine abnormalities constituting Albright syndrome (1,2–4,10–12,22). As more subtle forms of the bone pathology were appreciated, the monostotic form came to predominate in more recent series (9,14,19,20). Vertebral fibrous dysplasia is uncommon but not rare in polyostotic disease and increases in frequency with increasing involvement of the rest of the skeleton. Therefore, the early cases actually have fairly frequent vertebral involvement. Albright et al. (1) described five patients with severe polyostotic disease with the associated stigmata that now make up Albright syndrome. Three of these patients had vertebral involvement, one with L3 alone and the other two with multiple involvement. The patient of McCune and Bruch (12) had collapse of multiple vertebrae. The original article by Lichenstein (10) in 1938 stated that "lesions have noted roentgenographically on the vertebrae." One of the four polyostotic patients had vertebral involvement. Furst and Shapia (5), Albright and Reiferstern (2), Dockerty et al. (3), and Warrick (22) each reported on small

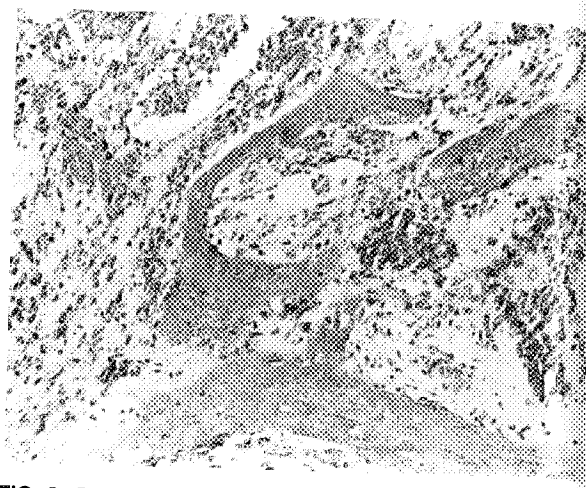

FIG. 3. Photomicrograph of the biopsy specimen reveals regular trabeculae of woven bone with intervening fibrous tissue typical of fibrous dysplasia.

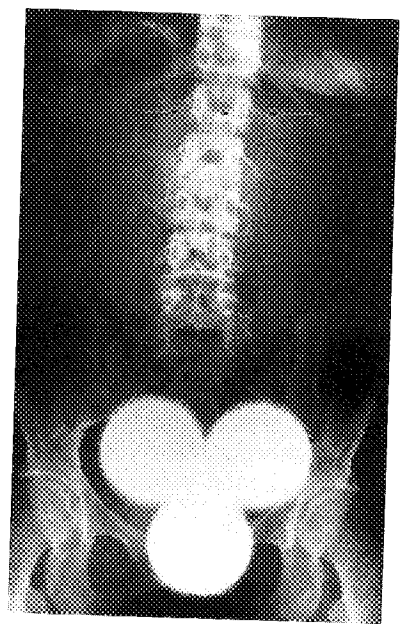

FIG. 4. Follow-up radiograph demonstrates fusion between L2 and L3. No scoliosis was apparent clinically.

numbers of patients, all with widespread polyostotic disease and a high incidence of vertebral involvement.

More recent series have been predominated by the monostotic form of the disease. Only two cases of monostotic fibrous dysplasia above the sacrum have been reported (8,20). Harris et al. (8) reported a series of 90 patients who presented over a 30-year period. Of the 50 cases that they described in detail, 37 were polyostotic (including those with Albright syndrome) and 13 were monostotic. They stated that of the polyostotic group, 14% had lumbar spine involvement and 7% had cervical spine involvement. Of the monostotic group, a single case involved the spine. The patient was a 42-year-old man who presented with low back pain. Radiographs revealed an expanded radiolucent lesion of a transverse process of L4. Biopsy confirmed the diagnosis of fibrous dysplasia. The single report of a monostotic lesion in the cervicothoracic spine is from Schlumberger (20). Of his 67 patients with monostotic fibrous dysplasia, one had a lesion of C4. The patient, a 20-year-old man, presented with 11 months of local tenderness and pain radiating down the arms. Seven months before the onset of symptoms, he had sustained a fracture of C5 without neurologic sequelae. Four months after the fracture (three months before the onset of the described symptoms), he sustained a blow to the same area. He apparently had a lesion of C4, which was biopsied. No radiographs or descrition of the distribution in C4 is provided. The initial pathological diagnosis was giant cell tumor, which was revised to fibrous dysplasia on review by the consulting pathologist.

Several recent reports refer only briefly to spinal involvement. Reed (16) reviewed 25 patients and found 16 with monostotic and nine with polyostotic fibrous dysplasia. Vertebral fibrous dysplasia (L2–L5) occurred in only one polyostotic patient and none in the monostotic group. Firat and Stutzman (4) had one case of sacral fibrous dysplasia in each of his their main groups: monostotic (n = 15) and polyostotic (n = 9). The series of Henry (9) of 50 monostotic patients had no vertebral involvement. The report by Schajowicz (19) of 222 monostotic cases and 36 polyostotic cases included a sacral lesion in the monostotic group.

The present case demonstrates monostotic fibrous dysplasia of the lumbar spine. A review of the major series of fibrous dysplasia indicates that this lesion is unusual. The single previously reported case of monostotic lumbar fibrous dysplasia involved only the transverse process. The present case involved the posterior elements but also extended to the vertebral body. Although the diagnosis was suspected preoperatively, the rarity of the presentation necessitated biopsy. The progression of individual lesions in fibrous dysplasia slows or halts at puberty (9,21). Malignant degeneration (13,17) and spinal cord compression (15) may occur; however, both have been reported only in severe polyostotic cases.

If the symptoms are minimal and the radiographic appearance is that of a benign lesion, surgical treatment is not mandatory. When there is diagnostic uncertainty, an excisional biopsy is indicated. Fusion is necessary only when removal of posterior elements has been extensive enough to produce instability.

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